Modern Concepts of Cardiovascular Disease

Published monthly by the AMERICAN HEART ASSOCIATION

44 EAST 23RD STREET, NEW YORK 10, N. Y.

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Vol. XXIV **SEPTEMBER 1955** **No. 9**

JONES CRITERIA (MODIFIED) FOR GUIDANCE IN THE DIAGNOSIS OF RHEUMATIC FEVER*

In 1944, the late Dr. T. Duckett Jones published criteria for the diagnosis of rheumatic fever which have been generally accepted in the United States and in many parts of the world. Subsequently Dr. Jones guided the revision of his criteria for use in the United Kingdom-United States Cooperative study on "The Relative Effectiveness of ACTH, Cortisone and Aspirin in the Treatment of Rheumatic Fever" and, just prior to his death, he participated in a conference on the revision of his original suggestions for use by the practicing physician. These modified Jones criteria are based in great measure upon his suggestions.

Rheumatic fever is related to previous infection with group A beta hemolytic streptococci, but the mechanism of the disease is unknown. Its boundaries are indefinite, and its differentiation from other diseases is sometimes impossible. There is no specific laboratory diagnostic test. The diagnosis must therefore be arbitrary and empirical. Criteria herein set forth are aimed at identifying those individuals who have had or are having an attack of rheumatic fever. They make no attempt to measure rheumatic activity at any given time or to diagnose inactive rheumatic heart disease. Thus, following the designation of an illness as rheumatic fever, the existence of continued activity or the presence of inactive rheumatic heart disease may be indicated by criteria different from those outlined below.

Criteria are necessary in order to minimize both overdiagnosis and underdiagnosis. The tendency to label as rheumatic fever a chronic febrile illness for which no obvious cause can be found is to be deplored. The tragedy which may lie in the wake of the false diagnosis of rheumatic fever may be even greater than the possible harm of missed recognition in questionable cases. The institution of effective prophylactic regimens requiring prolonged administration of sulfadiazine or antibiotic agents places a grave responsibility on the physician in the diagnosis of this illness.

In this statement, the diagnostic features of the disease are divided as originally proposed by Jones into major and minor categories dependent upon their relative occurrence in rheumatic fever and in other disease syndromes from which this disease must be differentiated. Thus chorea is included among the major criteria while fever, a symptom common to many diseases, is placed in a minor category. *These major and minor categories have no significance beyond their diagnostic import either as to prognosis, amount of "rheumatic activity," or severity of acute illness.* Indeed, a severe manifestation of rheumatic fever such as rheumatic pneumonia is not included because it is difficult to differentiate from congestive cardiac failure and because it almost always occurs in patients whose rheumatic fever is so obvious as to offer no difficulty in diagnosis.

The presence of two major criteria or one major and two minor criteria indicates a high probability of the presence of rheumatic fever (with one notable exception, see the last paragraph, page 294). In addition to the major and minor criteria to be used in the recommended formula, other manifestations have been listed which may be used to support the diagnosis. These criteria are not meant to substitute for the wisdom and judgment of the clinician. They are designed only to guide him toward a diagnosis of the disease with the suggestion that he follow carefully all questionable cases and restrict the diagnosis of rheumatic fever to illnesses which meet acceptable criteria.

*This is a report of the Committee on Standards and Criteria for Programs of Care of the Council of Rheumatic Fever of the American Heart Association and has been approved by the Executive Committee of that Council.

JONES CRITERIA (MODIFIED) FOR GUIDANCE IN THE DIAGNOSIS OF RHEUMATIC FEVER

Major Criteria

I. Carditis
II. Polyarthritis
III. Chorea
IV. Subcutaneous nodules
V. Erythema Marginatum

Minor Criteria

I. Fever
II. Arthralgia
III. Prolonged P-R Interval in the ECG
IV. Increased ESR, WBC, or presence of C-reactive protein
V. Preceding Beta-hemolytic streptococcal infection
VI. Previous rheumatic fever or inactive rheumatic heart disease

MAJOR DIAGNOSTIC CRITERIA

I. ***Carditis*** As evidenced by any one of the following:

A. The presence of a significant apical systolic murmur,[1] apical mid-diastolic murmur,[2] or basal diastolic murmur[3] in an individual without a history of previous rheumatic fever or in whom there is good reason to believe there was no pre-existing rheumatic heart disease; or a change in the character of any of these murmurs under observation in an individual with previous history of rheumatic fever or rheumatic heart disease.

B. Obviously increasing cardiac enlargement by x-ray.

C. Pericarditis manifested by a friction rub, pericardial effusion, or definite electrocardiographic evidence.

D. Congestive heart failure (in a child or young adult under 25) in the absence of other causes.

II. ***Polyarthritis*** Polyarthritis tends to be migratory and is manifested by pain *and* limitation of active motion, or by tenderness, heat, redness or swelling of two or more joints. Arthralgia alone without objective evidence of joint involvement is not a major manifestation.

III. ***Chorea*** This must be differentiated from habit spasm, athetosis, and cerebellar ataxia. Movements must be characteristic, involuntary and of moderate severity if chorea is to be used as a major manifestation.

IV. ***Subcutaneous Nodules*** Subcutaneous nodules are shot-like, hard bodies seen or felt over the extensor surface of certain joints, particularly elbows, knees and wrists, in the occipital region, or over the spinous processes of the thoracic and lumbar vertebrae.

V. ***Erythema Marginatum*** This recurrent, pink, characteristic rash of rheumatic fever in which the color gradually fades away from its sharp scalloped edge, is found mainly over the trunk, sometimes on the extremities, but not on the face. It is transient, is brought out by heat and migrates from place to place.

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[1] *A significant apical systolic murmur* is long, filling most of systole; is heard best at the apex; is as well transmitted toward the axilla as over the precordium; and does not change with position or respiration. It must be differentiated from an innocent (functional) murmur which is frequently found in normal people. This innocent murmur is systolic, occasionally harsh, is heard best along the left sternal border and usually changes with position and respiration. Borderline systolic murmurs, intermediate in location and nature, occur and should be carefully watched. Questionable murmurs which are intermittently present or which, after a period of observation, cannot be clearly classified as significant are rarely of any import.

[2] A significant organic *apical systolic murmur* is frequently accompanied by a low-pitched, short *mid-diastolic murmur* which is sharply localized to the chest wall over the apex of the heart and often heard best with a patient in the left lateral position with the breath held in expiration. This murmur, rarely present in the absence of an apical systolic murmur, confirms the significant nature of the latter. It must be differentiated from the long, low-pitched, crescendo apical presystolic murmur followed by an accentuated mitral first sound which is indicative of mitral stenosis but not of acute carditis.

[3] The development of a *basal diastolic murmur* of aortic insufficiency is also indicative of carditis. It is an early, short, diminuendo murmur usually heard only or heard best along the left sternal border in deep expiration. It has great diagnostic value, even though it may be difficult to hear and present only intermittently.

MINOR DIAGNOSTIC CRITERIA

I. ***Fever*** A significant rise in temperature is a common symptom, but, because it occurs in so many illnesses, it has little differential diagnostic value. In order to be included, the elevation in temperature must clearly exceed the normal diurnal fluctuation in which there is great individual variation.

II. ***Arthralgia*** Pain clearly located without objective findings is only a minor criterion for diagnosis. The pain must be in the joint, not in the muscles or other periarticular tissues, and must be distinguished from the nocturnal pain in the extremities occuring in normal children. Arthralgia must not be used as a minor criterion when polyarthritis is included as a major criterion.

III. ***Prolonged P-R Interval in the electrocardiogram*** Prolongation of the P-R interval may be nonspecific; it is considered a minor criterion and is not diagnostic of carditis. It cannot be used if carditis is already included as a major manifestation.

IV. ***Increased Erythrocyte Sedimentation Rate, Presence of C-reactive Protein, or Leukocytosis*** Elevation in one or more of these nonspecific tests may be considered as a single minor criterion. Particularly to be deplored is the tendency to use any of these tests as a major criterion or as diagnostic of rheumatic fever. There are many other nonspecific tests, but these three are most commonly used.

V. ***Evidence of Preceding Beta Hemolytic Streptococcal Infection*** This must be documented by (1) a history of scarlet fever or by a typical clinical picture of other streptococcal infection preceding the onset of rheumatic fever by one week to one month, the nature of the infection being confirmed by a history of immediate contact with other individuals having typical streptococcal infection or by positive culture of the nose or throat in which beta hemolytic streptococcus predominates; or (2) an elevated or rising antistreptolysin-O titer

VI. ***Previous History of Rheumatic Fever or the Presence of Inactive Rheumatic Heart Disease*** The existence of either of these may be used as a minor criterion to aid in deciding the rheumatic nature of the illness in question. For this use, the previous history must be documented by the same objective criteria as are set forth in this statement or by the presence of inactive rheumatic heart disease.

OTHER MANIFESTATIONS

These include systemic manifestations such as loss of weight, easy fatigability, elevated sleeping pulse rate (tachycardia out of proportion to fever), malaise, sweating, pallor or anemia, and local manifestations such as epistaxis, erythema nodosum, precordial pain, abdominal pain, headache, and vomiting. These as well as a family history of rheumatic fever, provide additional evidence of the presence of rheumatic fever but are not to be included as diagnostic criteria.

There are combinations of these diagnostic criteria which occur in the presence of other illnesses which must be ruled out before a definitive diagnosis is made. One combination in particular—polyarthritis, fever, and elevated sedimentation rate—is the weakest of all combinations of major and minor criteria. Diseases to be ruled out include rheumatoid arthritis, gonococcal arthritis, lupus erythematosus disseminatus, subacute bacterial endocarditis, nonspecific pericarditis with effusion, leukemia, sickle cell anemia, serum sickness (including manifestations of penicillin sensitivity), tuberculosis, poliomyelitis, undulant fever, and septicemias, particularly meningococcemia.

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